

Atypical and Complicated Kawasaki Disease in Infants Do We Need Criteria?

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Case reports suggest that infants with Kawasaki disease have atypical presentations and a high complication rate, likely related to delayed diagnosis and treatment. To date, no study of consecutive cases has compared infants with older children who have both atypical and typical Kawasaki disease. We retrospectively reviewed 44 cases of Kawasaki disease treated at our hospital from March 1980 to 1990: 11 (25%) were infants; 9 (20%) had atypical Kawasaki disease, of which 5 (56%) were infants; the male to female ratio was 1.7:1. Infants had a higher incidence of atypical Kawasaki disease (5 [45%] versus 4 [12%]; $P = .007$) and of coronary artery complications (7 [64%] versus 3 [9%]; $P = .002$), and coronary artery complications developed in all of the infants with atypical Kawasaki disease (5 [100%] versus 0 [0%]; $P < .01$). Yet, the other manifestations and laboratory changes were at least as common as in the older children. Coronary artery complications did not develop in any patient who received early intravenous immune globulin therapy. We suggest that in infants with Kawasaki disease, accepted criteria are too restrictive to allow early diagnosis and effective treatment. Until a definitive test is available, clinical judgment is required in the diagnosis of atypical Kawasaki disease. Intravenous immune globulin is known to be safe, and its early use in patients with suspected atypical Kawasaki disease is appropriate.

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Kawasaki disease is not new. It was first described in 1967 in Japan¹ and later in the English-language literature in 1974 by T. Kawasaki, who suggested the name mucocutaneous lymph node syndrome.² As early as 1959, however, in a review of 18 cases of infantile periarteritis nodosa, premortem and postmortem findings were noted to be strikingly similar to those of atypical Kawasaki disease.³ Infantile periarteritis nodosa and mucocutaneous lymph node syndrome have since been shown to be histologically indistinguishable.^{4,7} Atypical cases of Kawasaki disease are also histologically indistinguishable from the typical form,⁸ and the number of reports of this form of the disease has escalated in recent years.⁹

In a literature review of atypical Kawasaki disease, 23 case reports were found after 1980,⁹⁻²² and more are referenced in Japanese publications. In addition, a review of 45 atypical cases at the Japan Red Cross Medical Center was reported in 1987 (Japan Kawasaki Disease Research Committee, Department of Pediatrics, Japan Red Cross Medical Center, Tokyo). These authors have shown that atypical Kawasaki disease has the same,

but probably more frequent, occurrence of coronary artery complications and that it differs in some respects from typical Kawasaki disease. To date, there has not been a study of consecutive cases comparing infants with older children with both atypical and typical Kawasaki disease.

We report a retrospective review of 44 cases of Kawasaki disease seen consecutively at our hospital from 1980 to 1990 and compare the cases of infants with those of older children with both typical and atypical presentations. The study was directed at answering the following questions:

- Do infants differ from older children in manifestations or complications of Kawasaki disease?
- How does atypical Kawasaki disease differ from the typical form?
- Will any difference aid in early diagnosis and therapy?
- Can coronary artery complications and mortality be prevented?

ABBREVIATIONS USED IN TEXT

CDC = Centers for Disease Control and Prevention
IVIG = intravenous immune globulin

Patients and Methods

We reviewed the medical records of all children admitted to our hospital (Alberta Children's Hospital, Calgary) from January 1980 to February 1990 with a discharge diagnosis of Kawasaki disease or mucocutaneous lymph node syndrome. In addition, we reviewed all autopsy and echocardiogram reports from 1980 to 1990 at our hospital for the documentation of coronary artery aneurysm. The diagnosis of Kawasaki disease was accepted if one of the following conditions applied:

- Criteria set by the Centers for Disease Control and Prevention (CDC) were satisfied²³;
- Four of the CDC criteria were present, with the subsequent development of periungual desquamation and illness not explained by known pathogens; or
- At least two criteria were present, followed by the development of coronary artery aneurysm.

Patients who met the last two conditions were diagnosed as having atypical Kawasaki disease. All patients were seen by a pediatrician and, since 1985, by a pediatric infectious disease clinician. All patients also had some combination of viral serologic tests (including adenovirus, measles, Epstein-Barr virus), viral culture, and bacterial cultures of blood, urine, and cerebrospinal fluid.

Coronary artery complications, diagnosed in all cases by echocardiography, included coronary artery aneurysm or coronary artery dilatation. Coronary artery dilatation was defined as an arterial dilatation of greater than 2 mm in children younger than 2 years and greater than 2.5 mm in children older than 2 years.²² Coronary artery aneurysm was defined as the presence of an arterial dilatation greater than 4 mm.

Other associated manifestations included diarrhea consisting of at least four watery stools per day for at least one day; elevated liver function test values—either an aspartate or an alanine aminotransferase level of greater than 40 IU per liter; and aseptic meningitis—a cerebrospinal fluid leukocyte count of greater than 5×10^6 cells per liter (5 cells per mm³). Mucositis was considered to occur at three sites: pharynx, tongue, and lips; involvement of any one of these was enough to satisfy the criterion. An infant was defined as being 1 year old and younger.

Although the Fourth Revised Diagnostic Guideline from Japan (Japan Red Cross Medical Center) considers the presence of four criteria with coronary artery aneurysm as typical Kawasaki disease, this would diagnose cases at a stage too late for preventive intravenous immune globulin (IVIG) treatment, and we did not follow this guideline. Peripheral desquamation was not accepted as a criterion for typical Kawasaki disease because this also occurs late in the disease.

Statistical analysis of data was done using χ^2 and unpaired two-tailed Student's *t* test. Because of the multiple comparisons, significance was accepted at a *P* value of .01 or less.

Results

During the study period, 47 cases were identified with a discharge diagnosis of Kawasaki disease or mucocutaneous lymph node syndrome. No additional cases were identified on screening of all autopsy and echocardiogram reports. Of the 47 cases identified, 3 were excluded because they did not meet definitions for either typical or atypical Kawasaki disease, leaving 44 for analysis.

The patients ranged in age from 2 months to 9 years, 7 months (median, 25 months). There were 28 boys and 16 girls (male to female ratio, 1.7:1). Atypical Kawasaki disease accounted for 9 cases (20%). Infants accounted for 11 cases of Kawasaki disease (25%), with 5 cases of the atypical form (56%). Of the 5 infants with atypical Kawasaki disease, the cases of 3 (60%) satisfied four criteria, with subsequent periungual desquamation and illness not explained by known pathogens. All cases of atypical Kawasaki disease in infants satisfied at least two criteria, followed by the development of coronary artery aneurysm. Of the 35 patients with typical Kawasaki disease, 26 (74%) were younger than 4 years of age, 24 (68%) were younger than 3 years of age, and 17 (48%) were younger than 2 years of age. Fever was present in all 44 patients (100%).

Infants were found to differ from older children in several respects. Infants had substantially less extensive mucositis, pharyngitis, cervical adenopathy, and conjunctivitis. They also had a higher proportion with no mucositis (4/11 versus 0/33, *P* = .01). These differences were even more pronounced when infants with atypical Kawasaki disease were compared with older children (Table 1). Particularly noteworthy were the absence of cervical lymphadenopathy and the patchy mucosal involvement in infants with atypical Kawasaki disease. The most common findings were rash and fever.

The other manifestations and laboratory changes of Kawasaki disease were at least as pronounced in infants, including those with atypical Kawasaki disease, as in the older children. Infants had a high incidence of diarrhea and irritability, with a trend toward a more prolonged fever (*P* = .04, Table 1). Infants had elevated leukocyte counts, elevated platelet counts, and increased erythrocyte sedimentation rates, with a high incidence of sterile pyuria, elevated liver function test values, and a substantially lower hemoglobin level than older children (Table 1). Thus, despite fewer classic findings in the accepted criteria of Kawasaki disease, infants still had the other manifestations of the disease, including the laboratory changes.

Coronary artery complications were significantly more common in infants—7 (64%) versus 3 (9%) in children older than 1 year (*P* = .0002). This difference was most notable in infants with atypical Kawasaki dis-

TABLE 1.—Findings of Kawasaki Disease in Infants and in Older Children

Clinical Features	Kawasaki Disease					
	Atypical, ≤ 1 yr (n = 5)*		Typical, <1 yr (n = 6)*		All, >1 yr (n = 33)*	
	No.	(%)	No.	(%)	No.	(%)
Criteria						
Cervical lymphadenopathy	0	(0)	2	(33)	23	(70)††
Conjunctivitis	2	(40)	5	(83)	33	(100)††
Peripheral edema or erythema	2	(40)	4	(67)	25	(76)
Pharyngitis	2	(40)	2	(33)	27	(82)†
Diaper rash	1	(20)	4	(67)	18	(54)
Other rash	5	(100)	6	(100)	31	(94)
Mucositis ≤1 site ...	5	(100)	2	(33)	3	(9)††
Other symptoms						
Diarrhea	4	(80)	2	(33)	14	(42)
Irritability	5	(100)	5	(83)	24	(73)
Fever, days	15 ± 8.9		11.6 ± 5.7		9.6 ± 3.8	
Hospital days to diagnosis	4.0 ± 3.4		1.7 ± 2.4		1.7 ± 2.3	
Total days to diagnosis	8.8 ± 2.7		9.5 ± 4.0		7.9 ± 3.4	
Laboratory tests						
Leukocyte count, × 10 ⁹ /liter	30.5 ± 19		16.4 ± 3		17.5 ± 6	
Hemoglobin, grams/liter	80 ± 24		95 ± 5		104 ± 9†	
ESR, mm/hr	94 ± 13		89 ± 18		85 ± 26	
Platelets, × 10 ⁹ /liter	858 ± 326		971 ± 295		733 ± 196	
PMN in differential count, %	67 ± 12		63 ± 15		74 ± 10	
Sterile pyuria	4	(80)	3	(50)	25	(76)
Elevated liver enzymes	3	(60)	2	(33)	14	(42)
Male:female ratio ...	1.5:1		2:1		1.8:1	

ESR = erythrocyte sedimentation rate, PMN = polymorphonuclear leukocytes

*The number of infants and children affected (with percentages) is given; other values are given as n ± 1 standard deviation.

†P < .01 comparing infants ≤ 1 year with children > 1 year.

††P < .01 comparing infants ≤ 1 year with atypical Kawasaki disease with all children > 1 year.

ease, all 5 of whom had coronary artery complications (an incidence of 100%; $P < .01$). Overall, patients with atypical Kawasaki disease had a higher incidence of coronary artery complications—5 (56%) versus 4 (14%) in the typical cases ($P = .001$).

The benefit of IVIG therapy is evident in Table 2. Coronary artery complications did not develop in any patients who received IVIG treatment before echocardiographic evidence of such complications, regardless of age. In those not treated with IVIG, coronary artery complications developed in 7 infants (88%) and 3 children older than 1 year (25%). Coronary artery complications developed in all infants with atypical Kawasaki disease, and the only one in whom an aneurysm did not develop received IVIG therapy when coronary dilatation (diameter, 3.5 to 4 mm) was identified. The only death occurred in a 3-month-old infant who presented with atypical Kawasaki disease. The male to female ratio for coronary artery complications was 1.3:1 for infants; all

three of the children older than 1 year who had such complications were boys, and all had a coronary artery aneurysm.

Other findings of note include that desquamation of diaper rash occurred toward the end of the first week, whereas peripheral desquamation occurred toward the end of the second week. Irritability was present in 28 of the 30 children younger than 3 years (93%) and only 6 of the 14 children older than 3 years (43%; $P < .01$). Pneumonitis, diagnosed by evidence on chest x-ray film, occurred in 9 (26%) of the 35 children with typical Kawasaki disease and 2 (22%) of the 9 children with atypical Kawasaki disease. Gallbladder hydrosis was found in 2 cases (4%) on ultrasonography. On examination of the cerebrospinal fluid of 7 patients, 6 of whom were infants, 5 with atypical Kawasaki disease, aseptic meningitis was found in 4; all were infants, 3 with atypical Kawasaki disease (60%). A history of recent exposure to rug shampoo was reported in 10 cases (38% of all asked; $n = 26$).

Discussion

In 1959 Munro-Faure reported that most patients with infantile periarteritis nodosa had at some time during their illness at least six of the following symptoms and signs: fleeting macular skin eruptions, fever, leukocytosis, conjunctivitis, pharyngitis, cervical adenitis, and cough.⁶ Other reviews of infantile periarteritis nodosa from 1963 and 1968 also conclude that there is a rather constant clinical syndrome that in retrospect is identical to atypical Kawasaki disease.^{24,25} There is also a report of a nonfatal case of infantile periarteritis nodosa in which the diagnosis was based on angiographic evidence of coronary artery aneurysm that fits present-day criteria for atypical Kawasaki disease.²⁶ Munro-Faure suggested that

the persistence of these symptoms beyond the usual duration of an upper respiratory infection in an infant whose condition gives cause for concern, should lead to the consideration of necrotizing arteritis. . . . It may well be that early institution of therapy could avoid a fatal outcome.^{3(p925)}

Now, 36 years later, therapy with IVIG combined with high-dose salicylate is known to prevent coronary artery complications if given before symptoms

TABLE 2.—Incidence of Coronary Artery Complications (CAC) With and Without Intravenous Immune Globulin (IVIG) Therapy in Infants and Older Children With Kawasaki Disease

IVIG Therapy Given Before CAC Noted on Echocardiogram	Age, yr	Patients, No.	With CAC on Follow-up, No.	(%)
Yes	≤ 1	3	0	(0)
.....	> 1	21	0	(0)
No	≤ 1	8	7	(88)
.....	> 1	12	3	(25)*

*P < .01 using χ^2 .

develop.²⁷⁻³⁰ It is time to be concerned about the early diagnosis of atypical Kawasaki disease.

The concern of possibly missing the diagnosis of atypical Kawasaki disease in infants has been raised in several case reports.¹⁰⁻²² Echocardiography has been recommended for various indications: prolonged illness with thrombocytosis,¹⁶ fever lasting more than five days with rash,¹⁴ prolonged fever with subsequent desquamation,³⁰ prolonged fever with two other criteria,¹⁸ or two criteria with thrombocytosis.⁹ The most severe complication of Kawasaki disease is that of the coronary arteries, and if detected on echocardiogram, it should both support the diagnosis and lead to treatment. No evidence exists, however, that treatment with IVIG is effective if initiated after day 10 of illness.²⁸⁻³¹ Thus, by the time coronary artery complications develop—mean time to coronary artery dilatation, day 10²⁹—it may be too late for IVIG therapy to be effective. In fact, in our series, coronary artery complications were detected in four of ten patients, including three infants with atypical Kawasaki disease, by day 10 (days 6, 8, 9, and 9, respectively); in another three cases, complications developed on day 11, including in one infant with atypical Kawasaki disease. No coronary artery complications were seen with early IVIG therapy. Thus, a need for echocardiography, although important for diagnosis and follow-up, is inadequate as a criterion for early treatment, and a normal echocardiogram should certainly not preclude treatment.

Several reports have scored infants at higher risk for coronary artery aneurysm.^{32,33} The autopsy reviews of infantile periarteritis nodosa^{3,4,25} consist mainly of infants (>85%), supporting the view that they have more severe complications. In autopsy cases of Kawasaki disease, 54% are reported to be infants.⁵ In our series, the incidence of coronary artery complications was 14% in patients with typical Kawasaki disease, 33% in infants with typical Kawasaki disease, and 100% in infants with atypical Kawasaki disease. Even in patients with typical Kawasaki disease, there have been several attempts at predicting the risk for coronary artery complications.³²⁻³⁴ A good prediction has been reported with the modified ASAI score, using the single criterion of fever of 14 days or longer. These scores are all based on retrospective criteria, however, and do not aid in either diagnosing atypical Kawasaki disease or predicting the need for IVIG therapy early in the course of the disease. In infants with atypical Kawasaki disease, who are at greater risk for coronary artery complications, predictive scores also are not adequate for early diagnosis and treatment. In the absence of a definitive laboratory test or pathognomonic sign, clinical judgment must be relied on for the early diagnosis of atypical Kawasaki disease.

In this study, infants were compared with older children with Kawasaki disease to look for manifestations that may aid in early diagnosis. Although the number of infants was small—5 with atypical, 6 with typical Kawasaki disease—findings from our series of 44 patients are comparable to information available in the

literature (Table 3).^{3,4,9-22,35} For typical Kawasaki disease in infants, our results are similar to those of Burns and co-workers,¹¹ who found an even higher rate of coronary artery complication (71%) and mortality (14%) than in this study; cervical lymphadenopathy was the least common (43%) and rash the most common (100%) manifestation. For atypical Kawasaki disease in infants, our results are comparable to those found with infantile periarteritis nodosa^{3,4} and in case reports in the literature of atypical Kawasaki disease⁹⁻²²; the least common manifestation overall was lymphadenopathy, and the most common were fever and rash. The coronary artery complications rate was high in infants and particularly infants with atypical Kawasaki disease (86% to 100%); it was also high in atypical cases (90% in case reports and 56% in this series) relative to typical Kawasaki disease (20% in literature,²⁹ and 14% in this series). Most notable was the high mortality in atypical Kawasaki disease (11% to 35%), infants (0% to 14%), and infants with atypical Kawasaki disease (20% to 43%), compared with the generally accepted mortality in typical Kawasaki disease of less than 2%.^{29,32,36}

In our study, infants differed substantially from older children in several respects. Infants had fewer of the accepted criteria, including less mucositis, conjunctivitis, cervical adenopathy, and pharyngitis. Infants had a higher incidence of atypical Kawasaki disease (45% versus 12%, $P = .007$) and of coronary artery complications. Still, infants had a high incidence of the other manifestations of the disease, including the laboratory abnormalities. In infants with atypical Kawasaki disease, the same findings applied, with an even higher rate of coronary artery complications (of 100%) and the only death.

Other reports have also found infants to have a high incidence of the various manifestations suggested here as important in early diagnosis. Burns and associates report the cases of infants having pyuria (75%), aseptic meningitis (43%), respiratory signs (75%), and desquamation (75%), with increased erythrocyte sedimentation rates, thrombocytosis, leukocytosis, and anemia¹¹; they conclude that infants have an increased risk of coronary artery aneurysm and an atypical evolution of the disease. In a review of atypical Kawasaki disease, the authors comment on irritability (64%), diarrhea (29%), pyuria (44%), respiratory symptoms (47%), aseptic meningitis (75%), elevated aminotransferase levels (18%), and electrocardiographic changes (53%).⁹ The series on infantile periarteritis nodosa^{3,4,24,25} also refer to cough (50%), abnormal urinary findings (75%), and abnormal central nervous system signs including aseptic meningitis, abnormal electrocardiograms, leukocytosis (87%), anemia (70%), and diarrhea (65%).

We recognize that our study definitions may have created a bias for patients with atypical Kawasaki disease to have a high incidence of coronary artery complications. This high incidence (100%) in infants with atypical Kawasaki disease is not easily explained by this potential bias, however. The conditions defined for atyp-

TABLE 3.—Literature Data on Kawasaki Disease (KD)

Clinical Features	Children With Atypical KD			Infants With Typical KD		Infants With Atypical KD			
	Literature Case Reports, n = 20*†	Sonobe and Kawasaki, n = 45‡	This Series n = 9	Burns et al, n = 7§	This Series n = 6	IPN Review, 1959, n = 6	IPN Review 1977, n = 11¶	Literature Case Reports, n = 14*	This Series n = 5
Fever ≥5 days, %	90	73	89	86	100	83	91	93	100
Mucositis, %	50	76	78	100	100	67	18	36	60
Rash, %	55	40	100	100	100	100	81	72	100
Cervical nodes ≥1.5 cm, %	20	36	0	43	33	33	18	7.1	0
Peripheral extremity edema or erythema, %	15	80#	33	86	66	17	18	21	40
Conjunctivitis, %	45	78	55	86	83	67	36	43	40
≥3 Criteria, %	75	20	22	NA	NA	33	73	79	40
CAC, %	90	21**	56	71	33	100	100	86	100
Age, mo									
Mean	26	26	18.2±15.7	2.6	8.8	6.7	6.5	5.3	6.8
Median	7	NA	11	2	10	6	6	4.5	8
Male:female ratio	1:1	1.14:1	2.3:1	1.3:1	2:1	1.5:1	4.5:1	1:1	1.5:1
Age <1 year, %	70	<50	56	--	--	--	--	--	--
Mortality, %	35	NA	11	14	0	100	100	43	20

CAC = coronary artery complications, IPN = infantile periarthritis nodosa, NA = not available

*From Levy and Koren⁷; Ammerman et al¹⁰; Burns et al¹¹; Canter et al¹²; Cloney et al¹³; Friedman¹⁴; Fukushima et al¹⁵; Reller et al¹⁶; Rowley et al¹⁷; Schuh et al¹⁸; Avner et al¹⁹; Kleiman and Passo²⁰; Krapf et al²¹; and Sonobe and Kawasaki.²²†Of the 23 case reports in the literature, 3 were excluded, 2 (Ammerman et al¹⁰; Cloney et al¹³) because they satisfied 5 criteria and 1 (Levy and Koren⁷) because the diagnosis was based solely on "late-stage arteritic changes" in the renal, coronary, mesenteric, and meningeal arteries.‡From Sonobe and Kawasaki.²² In their series, atypical Kawasaki disease was defined as "3 or 4 of principal symptoms" with no clinical and laboratory evidence of any other disease known to mimic Kawasaki disease.§From Burns et al.¹¹||From Munro-Faure.³¶From Landing and Larson.⁴

#Includes desquamation as a peripheral extremity change.

**In Tada et al,³⁵ the percentage was 40%. In their series with ≤3 criteria, there was a 33% incidence of coronary artery complications.

ical Kawasaki disease were the same in infants and older children, yet none of the older children with atypical Kawasaki disease had coronary artery complications. In addition, three infants with atypical Kawasaki disease also met the definition of having four criteria with subsequent desquamation. The literature also supports the finding of high complication and mortality rates in these infants (Table 3). Finally, the purpose of our study was to identify findings in these patients, regardless of selection bias, that might be used to allow early diagnosis and treatment, thus preventing cardiac complications. Our findings may indeed apply only to more severely affected patients with atypical Kawasaki disease; however, this is the most important group to address.

With the small numbers, our results should be interpreted with some caution. Still, review of the literature (Table 3) elicited similar findings. Most important are the findings for atypical Kawasaki disease, because infants with typical Kawasaki disease do not pose as great a diagnostic dilemma as infants with atypical disease. Although prolonged fever, rash, and desquamation were common in patients with atypical Kawasaki disease, less reliance on other accepted criteria is appropriate for early diagnosis and treatment. More emphasis should be placed on such manifestations as irritability, diarrhea, hydrocele, and abnormal laboratory data (including leukocyte count, hemoglobin level, platelet count, erythrocyte sedimentation rate, sterile pyuria, liver function test values, and aseptic meningitis). We

suggest that in infants with Kawasaki disease, accepted criteria are too restrictive to allow early diagnosis and effective treatment. Until a definitive test is available, clinical judgment is required in the diagnosis of atypical Kawasaki disease. Intravenous immune globulin and high-dose salicylate therapy are known to be safe,^{27,28,37} and their early use in suspected cases of atypical Kawasaki disease is appropriate.

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